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PRINTED: 2001-12-04 13:11:41
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TITLE:	ORAL SURGERY, ORAL MEDICINE, AND ORAL PATHOLOGY
PUBLISHER/PLACE:	Mosby-Year Book St. Louis Mo
VOLUME/ISSUE/PAGES:	1984 Jun;57(6):616-9 616-9
DATE:	1984
AUTHOR OF ARTICLE:	Hall MB; Sclar AG; Gardner DF
TITLE OF ARTICLE:	Albright's syndrome with reactivation of fibrous d
ISSN:	0030-4220
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Albright's syndrome with reactivation of fibrous dysplasia secondary to pituitary adenoma and further complicated by osteogenic sarcoma

Report of a case

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To our knowledge, this is the only reported case of Albright's syndrome with reactivation of the fibrous dysplasia in adulthood, a marked elevation of growth hormone secondary to a pituitary adenoma, and development of osteosarcoma within the reactivated fibrous dysplasia. In addition, this patient also had hyperthyroidism with normal TSH levels, indicating the presence of two separate endocrinopathies. Reactivation of the fibrous dysplasia after years of arrested growth in this patient is good evidence that fibrous dysplasia does not "burn out" after adolescence but that growth ceases only when growth hormone declines to adult levels.
(ORAL SURG. 57:616-619, 1984)

Albright's syndrome was first described in 1937 as consisting of the triad of polyostotic fibrous dysplasia, melanotic pigmented patches on the skin (café au lait spots), and precocious menstruation.[1] Today it is known that this syndrome can be associated with a wide variety of endocrine disturbances, which can include sexual and somatic precocity, goiter and/or hyperthyroidism, hyperparathyroidism, Cushing's syndrome, and occasionally acromegaly.[2,3]

According to Lipson and Hsu,[2] only five well-documented cases of acromegaly in association with Albright's syndrome had ever been described prior to publication of their case in 1981.[2] In addition, very few cases of Albright's syndrome with well-documented pituitary lesions have been described.[2]

The skeletal lesions associated with Albright's syndrome usually appear early in life and run a variable clinical course.[4] While it is generally believed that the lesions become static after growth is complete, proliferation may continue after skeletal maturity has been attained.[5] Supporting this concept is the report of reactivation of quiescent lesions during pregnancy.[6] Accelerated growth in childhood with premature closure of the epiphyses often causes these patients to be below average height as adults.[7,8] While sarcomatous change in the skeletal lesions is not common, it has been estimated that the rate of malignant degeneration is 400 to 1,000 times the spontaneous rate for de novo bone sarcomas.[9]

Although many theories have been proposed, this syndrome with its related endocrine disorders remains one of unknown etiology and uncertain pathogenesis.[10] At present, there exist two prominent hypotheses concerning the mechanism of the reported endocrine disturbances. Associated endocrine disorders could result from a primary hypothalamic disorder causing excessive secretion of a variety of releasing hormones which result in subsequent pituitary overactivity and increased function of the target organs. Alternatively, DiGeorge,[11] suggests that autonomous endocrine gland hyperfunction, independent of the hypothalamic pituitary system, can occur in some patients and that this might be related to the multiple endocrine adenomatoses syndrome.

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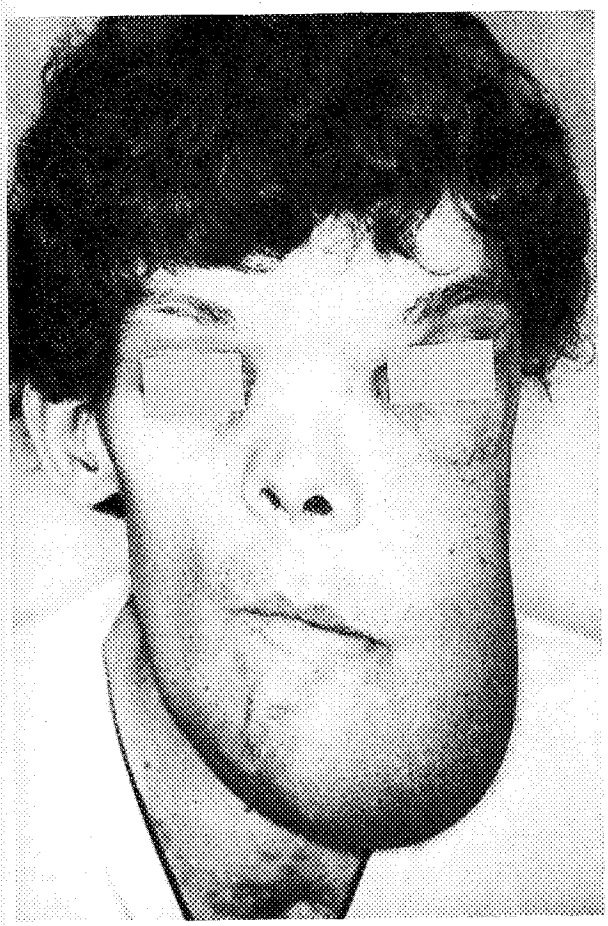

Fig. 1. Marked facial asymmetry of the left side of the face is evident in this photograph.

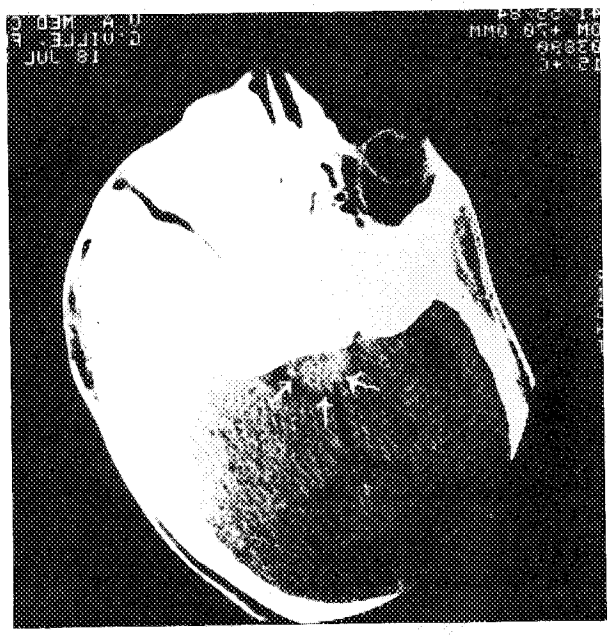

Fig. 2. Coronal CT scan demonstrating mass lesion in the sella region.

CASE REPORT

The patient, a white man, was 37 years old when admitted to the neurosurgery service on Aug. 4, 1980, for evaluation because of a recent complaint of headaches, growth consistent with acromegaly, and an outpatient growth hormone level of 69 ng/ml (normal, 0-5). The patient had a history of fibrous dysplasia, diagnosed at age 4, which resulted in marked distortion of the left face with frontal bossing, lateral displacement of the left orbit, and marked enlargement of the left maxilla and mandible. From 1960 to 1962, he underwent recontouring of the left maxilla and mandible, but repositioning of the left orbit was not attempted. In 1965 he underwent a partial thyroidectomy for toxic goiter, and in the same year he was found to have hypertension.

Facial growth was dormant until 1972; then the patient began noticing some subtle increase in size over the next five years. In 1977 there was enlargement of the remaining portion of the thyroid gland. Over the last 2 years prior to admission, the patient began to experience headaches, a gradual enlargement of the left side of the face, less distinct speech because of a tongue enlargement, the need to buy a large wedding ring, and inability to wear his dentures because of marked progressive enlargement of the left maxillary and mandibular alveolar ridges. Over the last few weeks he had noticed rapid enlargement of the left mandible and development of hypesthesia of the left lower lip.

Physical examination disclosed that he was 6 feet 8 inches tall. The left side of his face was markedly distorted with lateral displacement of the orbit and enlargement of the left forehead, left maxilla, and left mandible (Fig. 1). The right eye had a superior temporal quadrant defect, while the vision in the left eye was limited to light perception only. The left fundus showed optic atrophy, and the left ear canal was closed. The thyroid was enlarged on the right side, and there was evidence of the old thyroidectomy scar. Examination showed that the lungs, heart, abdomen, and nervous system were normal except for optic nerve atrophy with constant exotropia of the left eye, hypesthesia of the left lower lip and chin, and consistent tachycardia (pulse, 100). Hand films were consistent with acromegaly with mild tufting of distal phalanges.

Plain skull films and tomograms could not accurately define the sella turcica because of the massive increase in bone thickness of the calvarium. A CT scan showed a mass lesion in the sella region with 15 to 16 mm of suprasellar extension (Fig. 2). An x-ray film of the chest showed several enlarged right posterior ribs consistent with fibrous dysplasia. Thus, the fibrous dysplasia involved the bones of the left skull and face and several right ribs. The alkaline phosphatase was markedly elevated at 544 (normal, 30 to 120 IU/L), indicating evidence of increased bony remodeling. A thyroid scan showed an enlarged right thyroid with patchy uptake consistent with a nodular goiter. Thyroxine levels were increased, but thyroid-stimulating

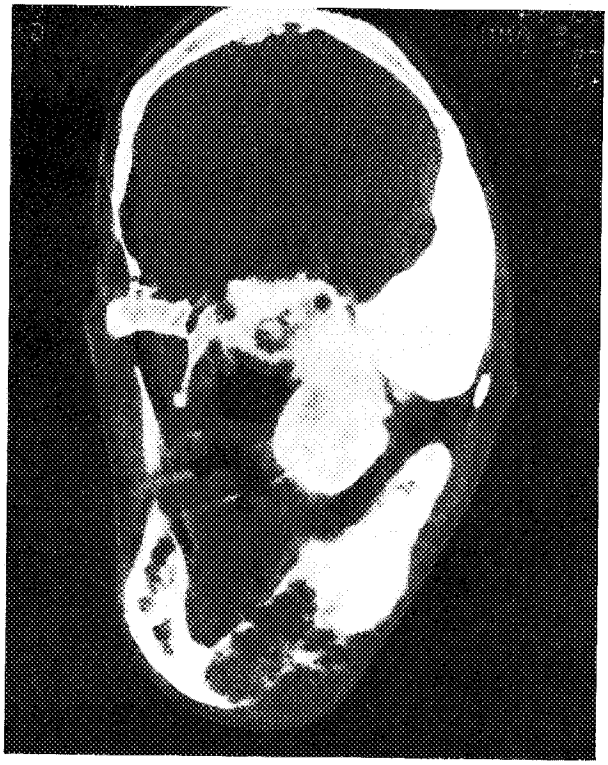

Fig. 3. Frontal CT scan demonstrating enlargement of left facial bones and radiolucency in left mandible indicative of osteosarcoma.

hormone (TSH) levels were not elevated. In addition, a thyroid-releasing hormone (TRH) stimulation test resulted in no detectable increase in TSH levels. Both of these last two tests indicated independent thyroid function.

An intraoral biopsy of the left maxilla and left mandible was performed. The maxillary biopsy was consistent with fibrous dysplasia. However, the mandibular biopsy revealed osteosarcoma within the fibrous dysplasia. A frontal CAT scan of the face and skull (Fig. 3) demonstrates the thickened left bones and a radiolucency in the left mandible representing the osteosarcoma.

Thus, this patient had (1) Albright's syndrome, (2) pituitary adenoma, (3) regrowth of fibrous dysplasia after at least 12 years of arrested growth, (4) primary hyperthyroidism, and (5) osteosarcoma in an area of previous fibrous dysplasia.

DISCUSSION

The primary reasons for reporting this case are (1) to add to the very small list of reported cases of Albright's syndrome with well-documented pituitary lesions, (2) to present a case of reactivation of fibrous dysplasia secondary to pituitary adenoma as evidence that these lesions have the ability to selectively regrow when stimulated by increased growth hormone, (3) to emphasize that osteosarcoma can develop in an area of fibrous dysplasia, and (4) to address the controversy regarding the underlying cause of the endocrinopathies associated with Albright's syndrome by presenting a case that supports the autonomous endocrine gland hyperfunction hypothesis.

In 1968 Firat and Stutzman[8] previously reported on this patient as demonstrating an unusual case of fibrous dysplasia. They came to the conclusion that he had polyostotic fibrous dysplasia, skin pigmentation, pituitary gigantism, and hyperthyroidism but noted that the sella was normal and made no report of growth hormone levels. We doubt the assumption of pituitary gigantism, since the family history of this patient is consistent with increased stature; his father is 6 feet 5 inches tall, his mother is 6 feet, and a sister has grown to 6 feet. We believe that the history of increased family stature and 12 or more years of quiescence of the bone lesions would suggest that the pituitary hyperactivity occurred with the development of a pituitary adenoma in adulthood.

We think it is plausible to suggest that the reactivation of the static lesions of fibrous dysplasia was due to the increased growth hormone levels associated with the pituitary adenoma. While reactivation has been reported during pregnancy, we did not find reports of reactivation secondary to pituitary adenoma.

In addition, this patient's clinical picture is unique in that it was further complicated by the formation of an osteosarcoma in the reactivated fibrous dysplasia of the left mandible. Finally, the low TSH levels noted in our patient and the fact that a TRH stimulation revealed no detectable increase in TSH levels indicate autonomous thyroid function and favor the hypothesis presented by DiGeorge.

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